

Timely Detection of Late - Onset Hearing Loss in Taiwan: Risk Factor Follow-up



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Helping Children with Hearing Loss Learn to Listen and Speak

OUTLINE

01

Background

02

Late-onset
hearing loss (HL) &
its impacts

03

Risk Factors

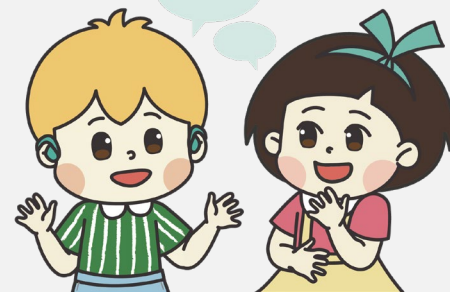
04

Follow-up timeline

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Conclusion

Leave your comments /
questions and contact info



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Background : In Taiwan



UNHS since 2012

2022: 98.7% screening rate



Preschool Hearing Screening

2021:

59.3% screening rate

10.3% referral rate



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Late-onset Hearing Loss

Definition

- Diagnosed after **3 months** of age in children who have passed UNHS



Prevalence

- 7.5 in 10,000 children who passed UNHS
- Accounts for 22% of children with bilateral HL

Challenges

- Parental difficulty in detection
- A pass in UNHS may foster the misconception that a child's hearing will always be normal



Aspects of Impact Due to Late-onset Hearing Loss in Children

- Ability to perceive sounds, words, and language rules
- Speech and language development
- Communication skills
- Cognitive development
- Social-emotional development
- Academic performance

Timely intervention is the key

to reducing the impact of late-onset hearing loss



Language
performance

Late-
onset HLL

=
N.S.

Early-
onset HLL

When intervention was provided, on average, 1 month
after identification for both groups

Risk Factor Monitoring: A Pivotal Approach to Timely Detection

- Regular monitoring of children who pass UNHS but exhibit risk factors for HL enables timely and effective detection of potential late-onset HL (Weichbold et al., 2006).



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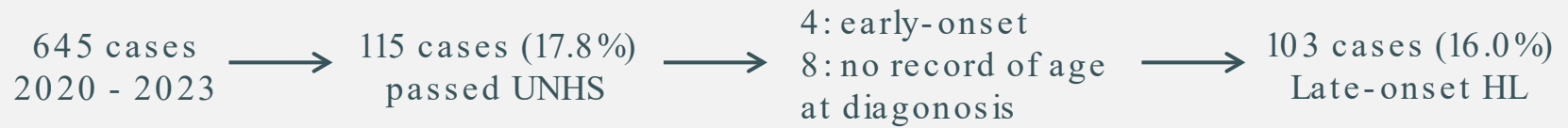
Risk Factors (1/2)

- **Perinatal factors**
 - Birth weight < 1500g
 - Apgar score 0-4 at 1min, 0-6 at 5min
 - NICU > 5 days
 - In utero infections: CMV, herpes, rubella, syphilis, toxoplasmosis
- **Medical treatments**
 - ECMO, assisted or mechanical ventilation, hyperbilirubinemia that requires exchange transfusion
 - Exposure to ototoxic medications (e.g., gentamicin, tobramycin) or loop diuretics (e.g., furosemide/Lasix)
 - Chemotherapy
- **Infections**
 - Culture-positive postnatal infections: bacterial and viral meningitis (especially herpes viruses and varicella)
 - Recurrent or persistent otitis media with effusion > 3 months

Risk Factors (2/2)

- **Family history & concerns**
 - Family history of permanent childhood hearing loss
 - Caregiver concern regarding hearing, speech, language, or developmental delay
- **Disorders**
 - Syndromes associated with hearing loss: neurofibromatosis type II, osteopetrosis, Usher, Waardenburg, Treacher Collins, Alport, Pendred, Jervell and Lange Nielson
 - Neurodegenerative disorders: Hunter syndrome, Friedreich ataxia, Charcot-Marie-Tooth syndrome
- **Craniofacial anomalies**
 - Anomalies including cleft lip / palate, microtia, aural atresia, choanal atresia, ear tags, ear pits, and temporal bone anomalies
 - Physical findings associated with syndromes known to include hearing loss (e.g., white forelock)
- **Head trauma**
 - especially basal skull/temporal bone fracture that requires hospitalization

Risk factor investigation for children in Children's Hearing Foundation (CHF)



Medical history review

Analysis of risk factors

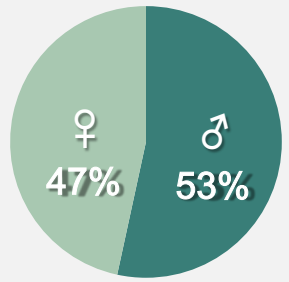
Descriptive statistics

may have had congenital HL undetected by UNHS



Demographics (N=103)

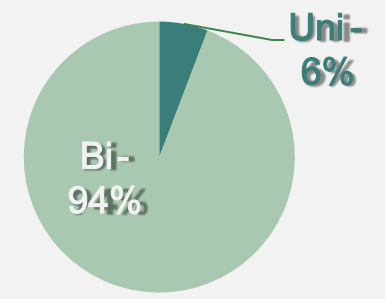
Sex



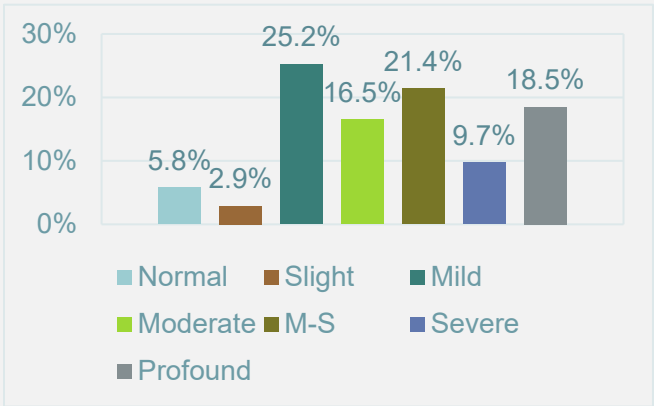
Age at diagnosis (mo.)

Mean: 39
SD= 26
Range: 4- 127

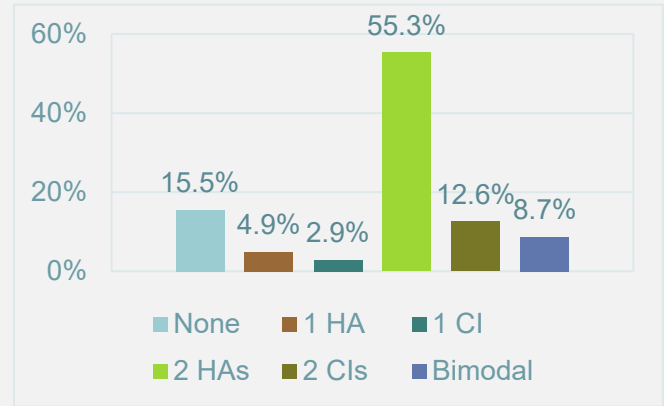
Uni- vs. Bi-lateral HL



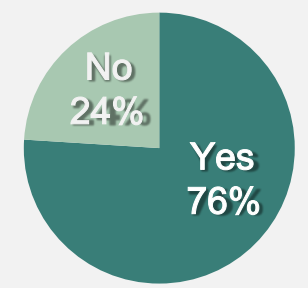
Degree of HL (better ear)



Technology



Intervention enrollment



Risk factors exhibited in CHF's children (1/2)

- **Perinatal factors**

- Birth weight < 1500g
- Apgar score 0-4 at 1min, 0-6 at 5min
- NICU > 5 days
- In utero infections: CMV, herpes, rubella, syphilis, toxoplasmosis

Gestational diabetes /
pre-eclampsia

- **Medical treatments**

- ECMO, assisted or mechanical ventilation, hyperbilirubinemia that requires exchange transfusion
- Exposure to ototoxic medications (e.g., gentamicin, tobramycin) or loop diuretics (e.g., furosemide/Lasix)
- Chemotherapy

- **Infections**

- Culture-positive postnatal infections: bacterial and viral meningitis (especially herpes viruses and varicella)
- Recurrent or persistent otitis media with effusion > 3 months

(Dumanch et al., 2017; J CIH, 1990, 2000, 2007, 2019; Baylan et al., 2010)

Risk factors exhibited in CHF's children (2/2)

- **Family history & concerns**

- Family history of permanent childhood hearing loss
- Caregiver concern regarding hearing, speech, language, or developmental delay

Identified HL
related gene(s)

- **Disorders**

- Syndromes associated with hearing loss: neurofibromatosis type II, osteopetrosis, Usher, Waardenburg, Treacher Collins, Alport, Pendred, Jervell and Lange Nielson
- Neurodegenerative disorders: Hunter ataxia, Huntington's disease, Friedreich's ataxia, Fragile X syndrome, Denton Tooth syndrome

Branchio-Oto-Renal
(BOR) Syndrome

Large vestibular
aqueduct syndrome
(LVAS) Syndrome

- **Craniofacial anomalies**

- Anomalies including cleft lip / palate, microtia, aural atresia, choanal atresia, ear tags, ear pits, and temporal bone anomalies
- Physical findings associated with syndromes known to include hearing loss (e.g., white forelock)

- **Head trauma**

- especially basal skull/temporal bone fracture that requires hospitalization

(Dumanch et al., 2017; J CIH, 1990, 2000, 2007, 2019)



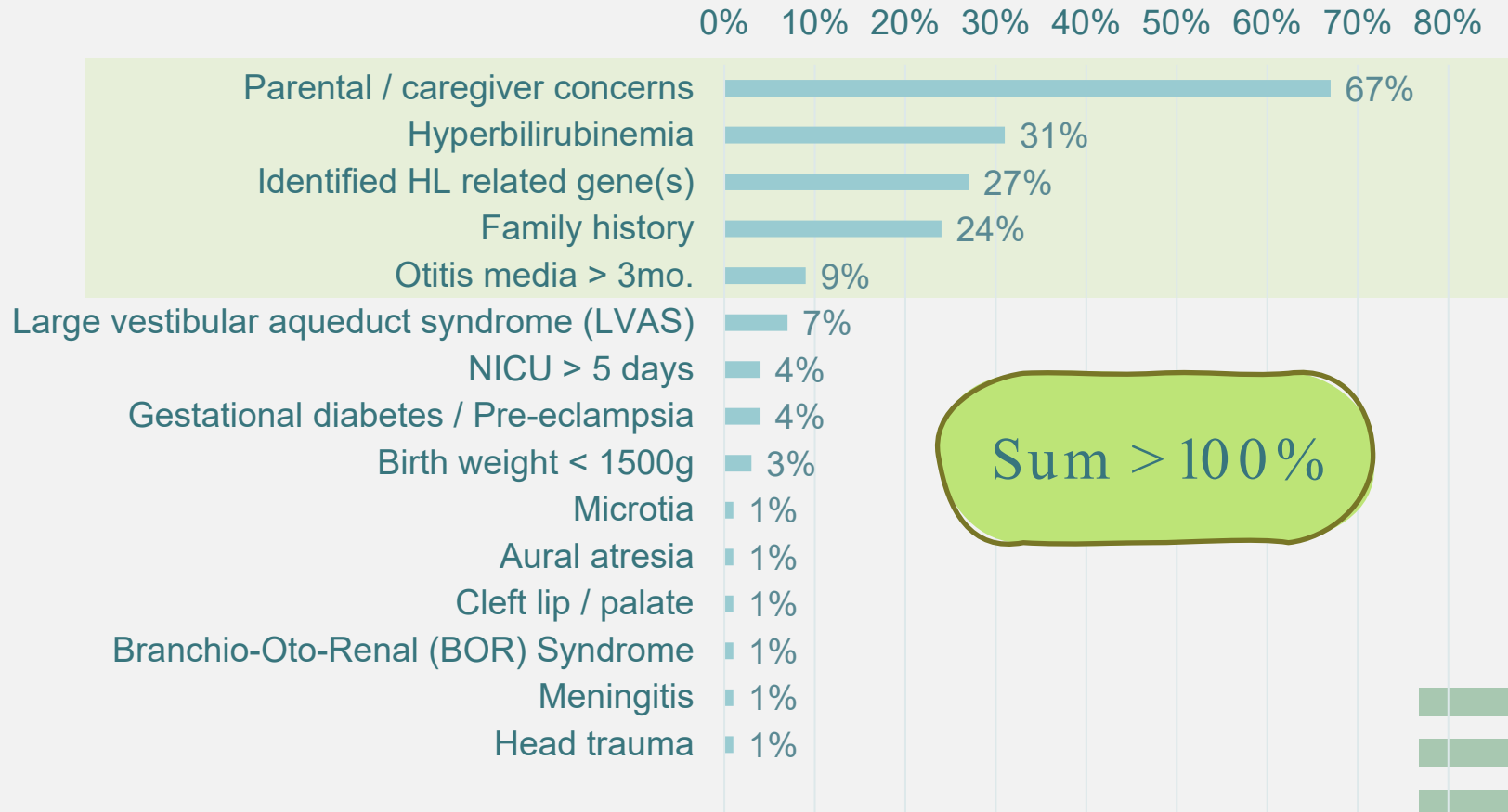
Take a guess!!

What were the 5 most prevalent risk factors among the 103 children with late-onset hearing loss in the CHF study?

Join at
slido.com
#3907 304



The proportion of risk factors present (N=103)



True examples of caregiver concern regarding hearing, speech, language, or developmental delay

At age 2, he remains non-verbal, sporadically producing meaningless sounds, and rarely vocalizing.

When she was younger, she responded well to calls, but later showed no response to calls from behind.

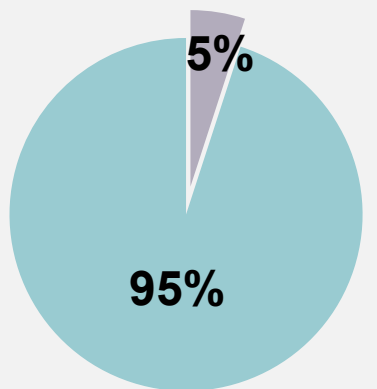
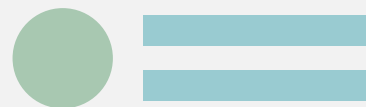
Currently, she only produces the sounds 'ma1' and 'ma4,' and has not yet produced 'ba4.'

Unable to form complete sentences, mostly using single words.

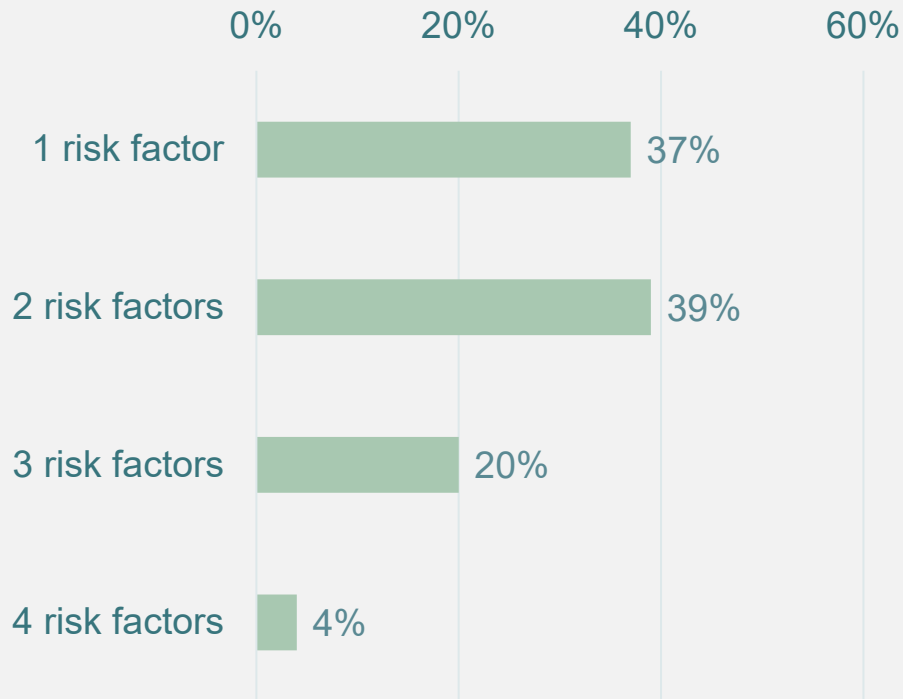
Displays poor response to sounds; slowly turns head to loud noises or rarely shows startle response even when being called loudly.



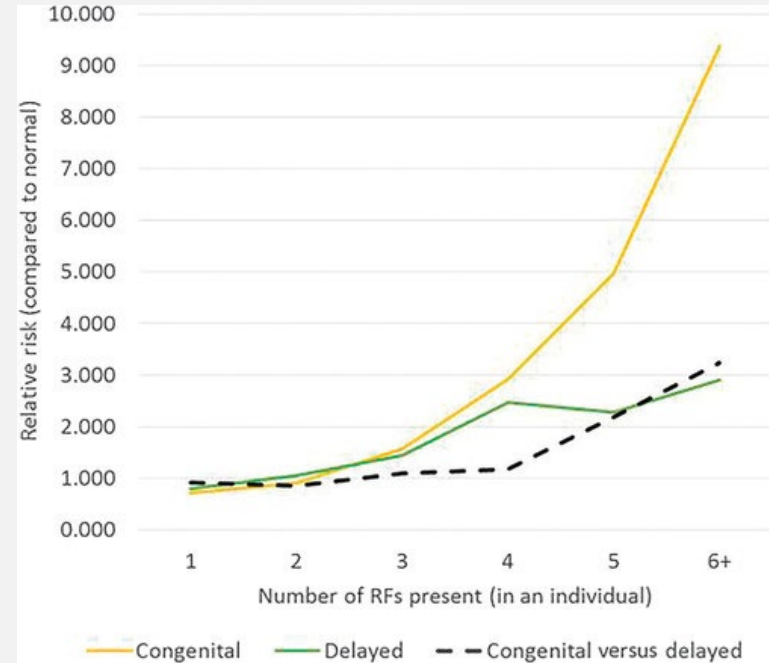
The occurrence of risk factors is variable and complex



■ No risk factors
■ One or more risk factors



The greater the number of risk factors exhibited, the higher the likelihood of both congenital and late-onset hearing loss



(Dumanchet al., 2017)

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Follow-up Timeline

Babies who passed UNHS

No risk factors

Ongoing surveillance of communicative development

Certain risk factors

- Caregiver concern
- Family history
- NICU > 5 days
- ECMO
- CMV
- Syndromes associated with HL
- Neurodegenerative disorders
- Meningitis
- Head trauma

Hearing evaluation by 6 months of age

Other risk factors

- Low birth weight
- Hyperbilirubinemia
- Ototoxic medications
- Congenital herpes, rubella, syphilis, toxoplasmosis
- Craniofacial anomalies
- Recurring or persistent otitis media with effusion > 3 mo.
- Others

Audiologic follow-up at 24-30 months of age

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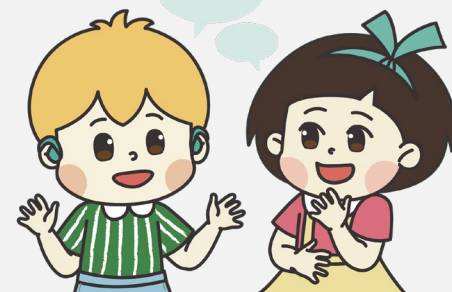
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Supporting children with late - onest hearing loss

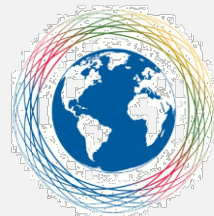
Risk factor follow-up

- Analysis based on CHF's database
- Analysis based on Taiwan Universal Health Insurance Database
- Public awareness promotion



Hearing performance checklist

- For children between ages of 3 and 6
- Parents and/or preschool/ kindergarten teachers
- Under development



2024
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VIRTUAL SYMPOSIUM
JUNE 26-27, 2024

Preschool hearing screening

- Public health nurse training for preschool hearing screening
- Advocacy for local government implementation of preschool hearing screening

Take home messages

- Reviewing and documenting the presence of **risk factors** for hearing loss is crucial in identifying the potential late-onset HL.
- As per J CIH guidelines, children who pass UNHS but present risk factors for hearing loss should undergo hearing evaluation by **6 months of age** , or **between 24 and 30 months of age** , depending on the specific risk factors.
- Even with negative results of UNHS, hearing and speech professionals must NOT overlook **caregiver concerns** regarding their child's hearing, speech, and language development.
- It is important NOT to disregard the possibility of hearing loss even in children **without identified risk factors** .





Acknowledgements

Coauthors

Social workers

Audiologists

AVTs

THANK YOU

Any questions?

Contact: yipingchang@chfn.org.tw



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comments
/ questions
and contact
info

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